Mediastinal and Retroperitoneal Teratoma with Focal Gastrointestinal Adenocarcinoma

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We report an unusual case of gastrointestinal adenocarcinoma arising in a giant posterior mediastinal mature cystic teratoma extending into the retroperitoneum, which was treated by complete excision with a good outcome for more than 2 years. Teratomas with malignant transformation are rare non-germ cell malignant tumors arising from a preexisting mature teratoma. Histological examination revealed that the cyst wall was composed of mature ectodermal, mesodermal, and endodermal elements. Neoplastic glands with a cribriform pattern were found in a small, solid nodule. Strong cytokeratin 20 cytoplasmic immunostaining of the tumor cells supported the diagnosis of gastrointestinal adenocarcinoma. In this report, we describe the potential aggressiveness of a giant mature cystic teratoma with adenocarcinoma and suggest that complete surgical resection without adjuvant chemotherapy be considered as a therapy in the treatment of teratoma with focal malignant transformation.

Key words: Transdiaphragm, Teratoma, Adenocarcinoma.

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Teratoma is defined as a tumor composed of elements from the mesoderm, endoderm, and ectoderm. Teratomas are found, in order of decreasing frequency, in the ovaries, testes, anterior mediastinum, and retroperitoneum.[1] Only a few cases have been reported to originate in the posterior mediastinum.[2] Teratoma with malignant transformation is extremely rare in gonadal, as well as extragonadal, sites. Squamous cell carcinoma accounts for 69% to 85% of the malignant transformation found in mature cystic teratoma of the ovary, and adenocarcinoma has been reported to occur in 6.8% of cases.[3–7] Most mediastinal teratomas occur in children and young adults. Almost 50% of patients remain asymptomatic. Older individuals more typically present with symptoms and signs related to intrathoracic compression, such as chest pain, cough, or dyspnea.[8]

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We report a rare case of gastrointestinal adenocarcinoma arising in a giant posterior mediastinal mature cystic teratoma extending into the retroperitoneum.

CASE REPORT

A 35-year-old man was admitted to the hospital because of low back pain lasting for 5 years and intermittent claudication. The pain radiated to bilateral lower limbs and was associated with numbness. A computed tomographic scan revealed a well defined, poorly enhanced soft tissue mass extending from the right posterior thoracic cage (Figure 1A) to the retroperitoneum with adhesion to the right psoas muscle (Figure 1B). A neurogenic tumor was favored. Tumor markers, including human chorionic gonadotropin, α-fetoprotein, carcinoembryonic antigen, and CA 19-9, were normal. No tumorous lesions were found in either testis. After obtaining informed consent, the patient was scheduled for an exploratory thoracotomy.

We entered the thoracic cavity through the 8th intercostal space by right thoracotomy with cutting posterior ends of the 8th to 10th ribs. The right diaphragm was then opened with resections of the 11th and 12th ribs for better exposure of the retroperitoneum. A large tumor measuring $20 \times 8 \times 8$ cm was located at the right lower posterior mediastinum with extension to the retroperitoneal space near the upper pole of the right kidney and adhesions to the aorta, lumbar spine, and right psoas muscle. Complete tumor excision was undertaken. The patient left the hospital on the eighth postoperative day.

The resected tumor weighed 400 g. The cut surface revealed multiloculated cystic cavities ranging from 0.5 to 2.5 cm. Most of them contained brown turbid material. The cyst lining was thickened with a yellow, gritty surface. From part of the cyst wall, an elastic firm solid nodule measuring $2.0 \times 1.4 \times 0.6$ cm was noted. Histologically, the cyst wall disclosed a mature cystic teratoma composed chiefly of mesodermal derivatives, such as adipose tissue, cartilage (Figure 2A), and smooth muscle, and endoderm characterized by gastrointestinal tract tissue (Figure 2B). Keratinizing squamous epithelium of ectoderm origin and calcification were occasionally seen. Sections from the solid nodule showed a neoplasm composed predominantly of tubular glands arranged in a cribriform pattern (Figure 3A). Non-neoplastic colonic epithelium was noted adjacent to the tumor. Immunohistochemical staining was negative for cytokeratin 7 but strongly positive for cytokeratin 20, supporting the diagnosis

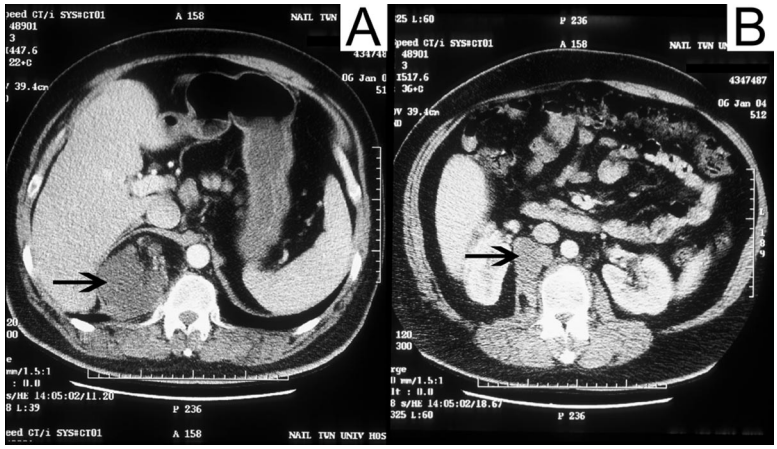

FIGURE 1. (*A*) Computed tomographic scan of the chest shows a large lobulated tumor (*arrow*) in the posterior mediastinum. (*B*) The tumor (*arrow*) protrudes through the right diaphragm to the retroperitoneum.

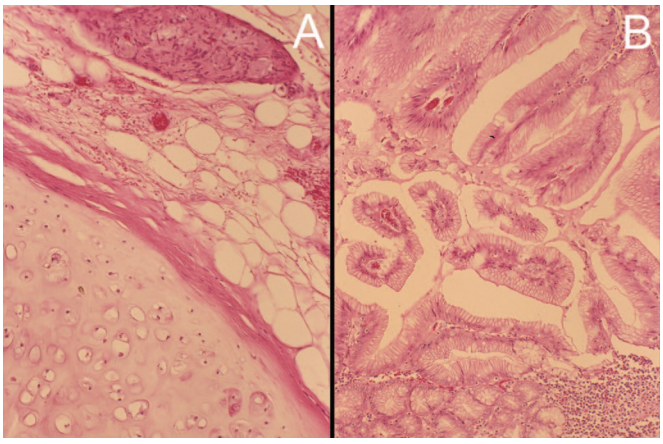

FIGURE 2. The mature cystic teratoma is composed of (*A*) cartilage and (*B*) gastrointestinal tract tissue. Hematoxylin-eosin staining; original magnification ×33.

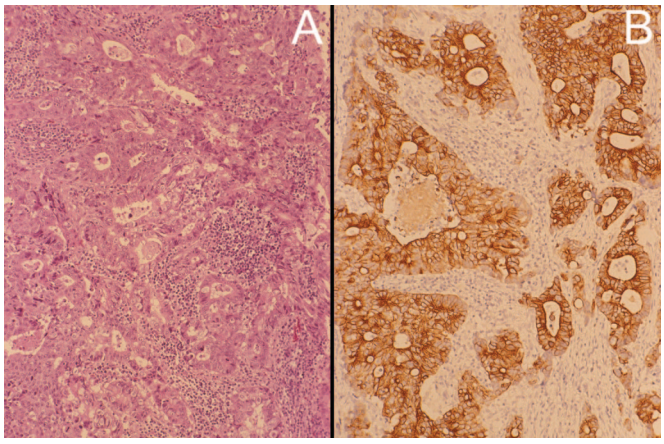

FIGURE 3. (*A*) The solid nodule of the mature teratoma demonstrates a well to moderately differentiated gastrointestinal-type adenocarcinoma composed of hyperchromatic cancer cells in tubular or cribriform arrangements. Hematoxylin-eosin staining; original magnification ×33. (*B*) Strongly CK20 immunoreactive neoplastic glands. ABC method; original magnification ×33.

of gastrointestinal adenocarcinoma (Figure 3B). There were no immature teratomatous elements or other germ cell components.

Because there were no signs of dissemination of the disease, no additional therapy was applied. The patient has been free of disease for more than 2 years.

DISCUSSION

The presence of non-germ cell malignancies within germ cell neoplasms is classified as teratoma with malignant transformation (TMT), according to the World Health Organization.[9] TMT is extremely rare, and most are found in gonadal tumors from the ovary[7] and the testis.[10] It occurs in 1% to 2% of mature cystic teratomas examined and comprises 2% to 3% of testicular teratomas. TMT of extragonadal origins is even rarer and has been described only in the anterior mediastinum,[3] stomach, brain, sacrococcygeal region, and retroperitoneum.[1] To the best of our knowledge, this is the first case of a large TMT with gastrointestinal adenocarcinoma of posterior mediastinum with retroperitoneal extension. Characteristically, TMTs are seen more frequently than mature teratomas in older patients (with a peak incidence in the fifth and sixth decades of life), and usually with a long-standing mature teratoma.[10] The most common malignant transformation is squamous cell carcinoma (69-85%), followed by adenocarcinoma and carcinoid.[3–7] Some reports suggest that the malignant transformation occurs from preexisting mature teratomatous elements.[1,3,10] The exact etiology responsible for the transformation remains to be determined. However, we believe that the risk of TMT may have increased because of the long-standing mature teratoma. Adenocarcinoma of uncertain origin is a frequent clinical problem, the distinction of which has a significant therapeutic and prognostic guide in TMT. The utility of cytokeratin immunostaining is helpful for the differential diagnosis. Adenocarcinomas arising from the gastrointestinal tract are generally positive for CK20 but negative for CK7.[10] Postoperative prognosis is good if the tumor is completely excised.[1,3]

In conclusion, complete resection of teratoma is justified if the tumor has not extended beyond the capsule because of the minimal risk of malignant transformation.

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